Case Report

Septic shock due to infected giant venous malformation complicated by massive bleeding

Keisuke Tomita,[1] Eizo Watanabe,[1] Tomohito Sadahiro,[1] Yoshihisa Tateishi,[1] Koichiro Shinozaki,[1] Naoki Rikihisa,[2] and Shigeto Oda[1]

[1]*Department of Emergency and Critical Care Medicine, Graduate School of Medicine, Chiba University, and* [2]*Department of Plastic, Reconstructive, and Esthetic Surgery, Chiba University Hospital, Chiba City, Japan*

Case: A 19-year-old man with a hemangioma that extended from the left arm to the axillary region had deteriorated due to shock, and no improvement was observed after fluid resuscitation. His status on arrival led to complications of hemorrhagic and septic shock with his left arm swollen and deep purple in color. Left arm amputation to control the source of bleeding and infection was thought to be indicated, however, the coagulation system had collapsed. Therefore, initial reduction of the blood flow to the hemangioma with angiographic endovascular treatment was carried out. Thereafter, continuous hemodiafiltration using a polymethylmethacrylate membrane hemofilter against hypercytokinemia was introduced in the intensive care unit for damage control, which resulted in success.

Outcome: We completed the arm amputation, and subsequently carried out a latissimus dorsi muscle flap transfer on the amputated stump. The patient achieved an ambulatory discharge.

Conclusions: We successfully treated the very rare case of massive venous malformation with shock due to hemorrhage and infection by performing damage control.

Key words: Disseminated intravascular coagulation, hemangioma, hemorrhagic shock, Klippel–Trenaunay syndrome, localized intravascular coagulopathy

INTRODUCTION

VENOUS MALFORMATION (VM) is sometimes accompanied by thrombocytopenia due to mass consumption of coagulation factors in the malformation vessels, hemolytic anemia, and the coagulation abnormality known as localized intravascular coagulopathy.[1] Here, we report the case of a patient who developed septic shock after a localized infection of a giant VM, which progressed to hemorrhagic shock due to bleeding from the infection focus. We successfully treated the patient with a combination of surgery and endovascular repair as damage control and intensive care to improve the patient's general condition.

Corresponding: Eizo Watanabe, MD, PhD, Department of Emergency and Critical Care Medicine, Graduate School of Medicine, Chiba University, 1-8-1 Inohana, Chuo-ku, Chiba City, Chiba 260-8670, Japan. E-mail: watanabee@faculty.chiba-u.jp.
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CASE

A 19-YEAR-OLD MAN HAD been diagnosed during childhood with a hemangioma that extended from the left arm to the axillary region. He developed nausea, vomiting, and fever (body temperature, 41°C), and experienced cold sensations in his limbs, along with considerable swelling on his left arm. At the time of consultation with his local physician, he was in a state of shock, and no improvement in hemodynamics was observed after fluid infusion and blood transfusion. Hence, he was referred to our department, and his level of consciousness was E1VTM1 according to the Glasgow Coma Scale. His vital signs were: body temperature, 38.1°C; heart rate, 150 b.p.m.; systolic blood pressure, 62 mmHg; and respiratory rate, 30 breaths/min. Although no external hemorrhage was observed, part of the left arm was remarkably swollen and deep purple in color (Fig. 1A).

Laboratory findings

The patient presented with a hemoglobin level of 6.2 g/dL and anemia due to hemorrhage into soft tissues, a decreased

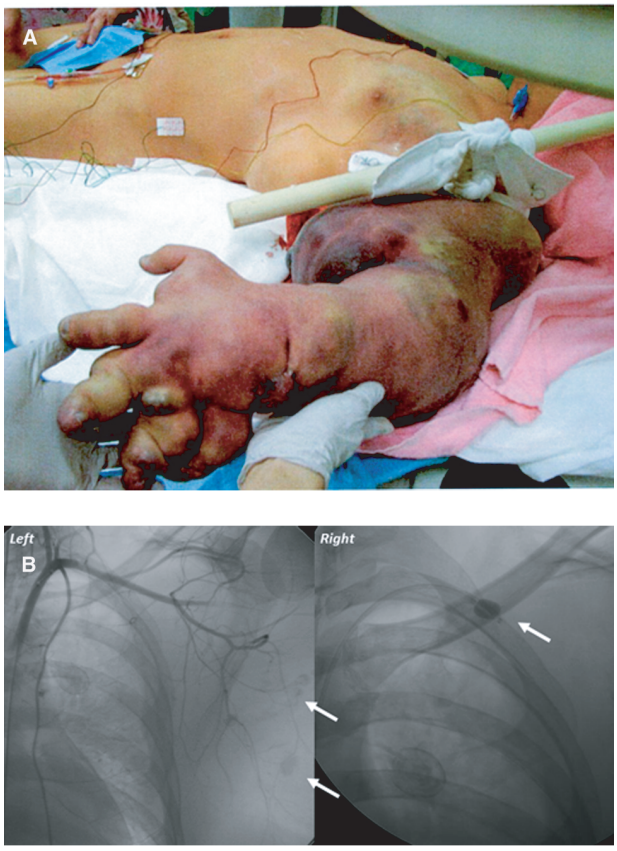

Fig. 1. Hemorrhage from an infected giant venous malformation in a 19-year-old man. A, Hemangioma in the thorax and left upper limb. The left upper limb was remarkably swollen and dark violet in color. B, Angiographic image before the first operation. Left, Multiple extravasation but otherwise unremarkable (white arrows). Right, inflated balloon (white arrow).

blood platelet count of 24,000/mm^3, an elevated fibrin degradation product level of 35.8 μg/mL, an unmeasurable prothrombin time activity, and prominent abnormalities of the coagulation/fibrinogenolysis system, leading to a diagnosis of disseminated intravascular coagulation.[2] The leukocyte count was slightly reduced to 3,300/mm^3, the serum interleukin-6 (IL-6) concentration was 182,000 pg/mL, and the serum lactate concentration was considerably elevated to 8.8 mmol/L, leading to a diagnosis of complications of hemorrhagic shock with septic shock.[3] In addition, blood urea nitrogen and creatinine levels were elevated to 49 and 4.67 mg/dL, respectively. The Acute Physiology and Chronic Health Evaluation II score was 34. On the following day, a blood culture carried out on admission revealed positives for *Streptococcus dysgalactiae* subspecies *equisimilis,* a group G hemolytic streptococcus.

Clinical course

On admission, the patient's response to initial resuscitation with a massive blood transfusion was poor. Although we administered 6 units of red cell concentrates rapidly, the hemoglobin level elevated only to 7.5 g/dL and he was still suffering from shock (heart rate, 150 bp.pm.; systolic blood pressure, 62 mmHg). We therefore considered carrying out left arm amputation to control the source of the bleeding and infection. However, we assumed that this would make achieving bleeding control difficult because of a coagulation collapse. Therefore, we decided to initially reduce the blood flow to the hemangioma using balloon occlusion. Angiography revealed dense staining of the hemangioma but no remarkable extravasation. A balloon catheter was then used to occlude the left subclavian artery (Fig. 1B)—the feeding artery of the hemangioma of the left arm—after which we attempted to amputate the left arm. However, a coagulation disorder led to massive intraoperative bleeding that was difficult to control. The bleeding point from the VM in the s.c. region of the left side of the chest moved deep into the s.c. pocket (15 cm from the skin incision) owing to the rupture of the spongy venous structure. The narrow field of view made hemostasis extremely difficult. We abandoned the left arm amputation to adopt a damage control strategy. Postoperatively, while in the intensive care unit (ICU), the patient was hemodynamically unstable. In addition to catecholamine supports, low-dose steroids were given for relative adrenal insufficiency.[4] A high dose of penicillin was already administered as an antimicrobial to target the group G streptococcus detected in the blood culture. Combination therapy of meropenem, linezolid, and clindamycin was also started empirically. Furthermore, continuous hemodiafiltration by using a polymethylmethacrylate membrane hemofilter (PMMA-CHDF) was initiated to address the acute kidney injury and hypercytokinemia.[5] The patient's hemodynamics stabilized with decreasing blood level of IL-6 through cytokine modulation by PMMA-CHDF (IL-6 on admission, 18,200 pg/mL; on day 1 after CHDF initiation, 56,500 pg/mL; on day 2, 3,070 pg/mL; on day 3, 582 pg/mL). On day 4 after ICU admission, we were able to carry out the amputation of the left arm. Microscopic findings of the resected hemangioma specimen revealed the growth of a cavernous hemangioma with dilated blood vessels and phagocytosis of Gram-positive cocci by white blood cell aggregates (Fig. 2A, B). On day 7 after ICU admission, the gauze that had been packed into the s.c. pocket was removed, and the wound was closed. On day 22, his renal failure and inflammation improved, enabling discontinuation of the PMMA-CHDF. On day 25, he was discharged from the ICU. Subsequently, a latissimus dorsi muscle flap transfer was carried out on the amputated stump, and sclerotherapy was

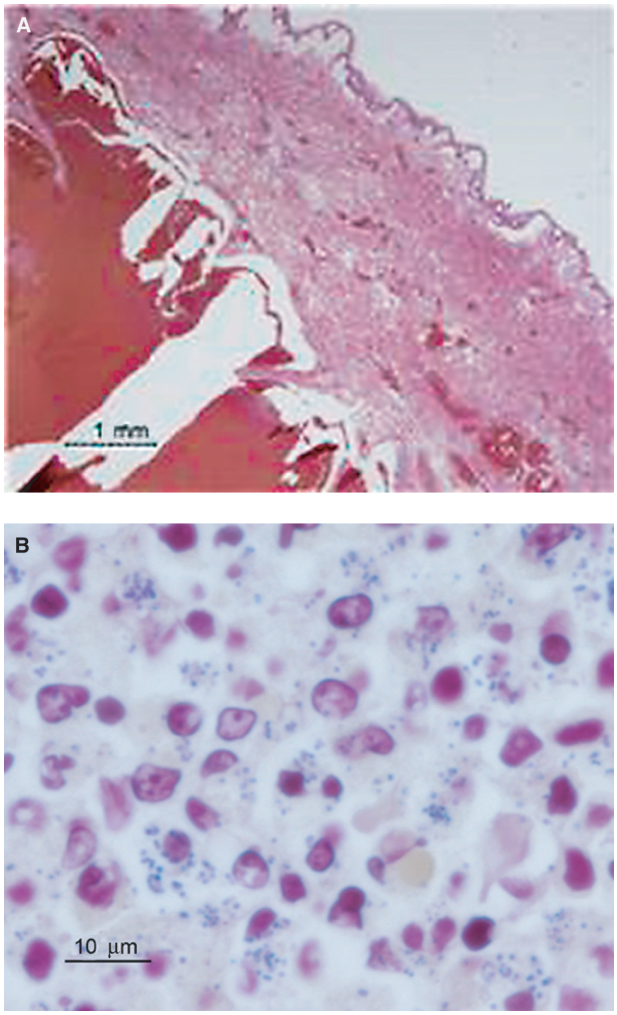

Fig. 2. Microscopic examination of a resected hemangioma specimen from the amputated left arm of a 19-year-old man with infected giant venous malformation. A, Multiple vascular cavities were observed (hematoxylin–eosin staining). B, Accumulation of leukocyte-phagocytizing Gram-positive cocci was observed (Gram staining).

carried out in stages to treat the thoracic wall venous malformations. On hospital day 84, the patient was given an ambulatory discharge.

DISCUSSION

HEMANGIOMAS AND VASCULAR malformations on the body surface had been commonly referred to as "hemangiomas." However, according to the International Society for the Study of Vascular Anomalies' classification system, which has become the global standard since its adoption in 1996, vascular malformations are now divided into two broad categories, namely neoplastic lesions and congenital malformations. The classification system acknowledges both disease concepts and their respective treatment methods.[1,6] In the present patient, VM was the main symptom, a condition that is conventionally referred to as "cavernous hemangioma." Furthermore, the patient was diagnosed with Klippel–Trenaunay syndrome on the basis of an overgrowth of the affected limb (bone and soft tissues). Patients with this syndrome present with an overgrowth of soft tissues in the affected limb, mesodermal abnormalities associated with vascular malformation, and decreased blood flow. It is generally considered to be a non-inheritable disease.[7] The condition is characterized by three features, namely port-wine stains, congenital abnormalities of venous formation, and an enlarged affected limb due to an overgrowth of the bone and soft tissues.[8]

Giant VMs are sometimes accompanied by a generalized coagulation disorder, decreased fibrinogen or platelet count, or elevated D-dimer. This was suggested to be due to the mass consumption of coagulation factors as localized intravascular coagulopathy and differs from the Kasabach–Merritt phenomenon, in which the patient presents with mass consumption of blood platelets in vascular tumors.[1] Some reports indicate the validity of transcatheter arterial embolization (TAE) for symptom improvement and as preoperative treatment of localized arteriovenous malformation with symptoms.[9] As for this case, we could not control the massive bleeding by TAE alone, due to both coagulopathy associated with severe infection and hemorrhage from multiple feeding arteries into the VM. Hence, we added gauze packing to achieve temporary hemostasis from both VM and feeding arteries. The intraoperative bleeding had been diminished by the TAE, which was thought to make temporary hemostasis easier. Because left arm amputation in a single surgical session was deemed difficult as a consequence, we aimed to stabilize the patient's general condition in the ICU by performing "damage control."

Klippel–Trenaunay syndrome is estimated to affect at least 1 in 100,000 people worldwide (http://ghr.nlm.nih.gov/condition/klippel-trenaunay-syndrome, accessed 8 July, 2015), however, the present case should be considered extremely rare because our search of published work did not reveal any other similar reports in which extensive VM and incredibly unstable hemodynamics resulting from concurrent hypovolemic shock and septic shock were reported.

CONFLICT OF INTEREST

NONE.

REFERENCES

1 Enjolras O, Wassef M, Chapot R. Color Atlas of Vascular Tumors and Vascular Malformations. New York: Cambridge University Press, 2007.
2 Gando S, Iba T, Eguchi Y *et al*. A multicenter, prospective validation of disseminated intravascular coagulation diagnostic criteria for critically ill patients: Comparing current criteria. Crit. Care Med. 2006; 34: 625–31.
3 Oda S, Hirasawa H, Shiga H, Nakanishi K, Matsuda K, Nakamura M. Sequential measurement of IL-6 blood levels in patients with systemic inflammatory response syndrome (SIRS)/sepsis. Cytokine 2005; 29: 169–75.
4 Annane D, Sebille V, Charpentier C *et al*. Effect of treatment with low doses of hydrocortisone and fludrocortisones on mortality in patients with septic shock. JAMA 2002; 288: 862–71.
5 Hirasawa H, Oda S, Nakamura M, Watanabe E, Shiga H, Matsuda K. Continuous hemodiafiltration with a cytokine-absorbing hemofilter for sepsis. Blood Purif. 2012; 34: 164–70.
6 Enjolras O. Classification and management of the various superficial vascular anomalies: Hemangiomas and vascular malformations. J. Dermatol. 1997; 24: 701–10.
7 Oduber CE, van der Horst CM, Hennekam RC. Klippel–Trenaunay syndrome: Diagnostic criteria and hypothesis on etiology. Ann. Plast. Surg. 2008; 60: 217–23.
8 Redondo P, Aguado L, Martínez-Cuesta A. Diagnosis and management of extensive vascular malformations of the lower limb: Part I. Clinical diagnosis. J. Am. Acad. Dermatol. 2011; 65: 893–906.
9 Woodhams R, Ogasawara G, Ishida K *et al*. Successful treatment of acquired arterial venous malformation using N-butyl-2-cyanoacrylate under balloon occlusion. Acta Radiol. Short Rep. 2014; 3: 1–6.